

Primary Sclerosing Cholangitis with Dilation of a Dominant Stenosis



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Abstract

The majority of patients with primary sclerosing cholangitis (PSC) develop stenoses of major bile ducts. Stenoses may be recognized by magnetic resonance cholangiopancreatography, but for definite diagnosis and especially for therapeutic decisions, endoscopic retrograde cholangiography still represents the gold standard. Repeated balloon dilations allow opening of total or subtotal strictures of the large bile ducts, the so-called dominant stenoses. A case with PSC admitted for endoscopic treatment of dominant stenoses is demonstrated. This article is part of an expert video encyclopedia.

Keywords

Balloon dilation; Cholangiocellular carcinoma; Cholangitis; Dominant stenoses; Endoscopic retrograde cholangiopancreatography; Jaundice; Video.

Video Related to this Article

Video available to view or download at doi:10.1016/S2212-0971(13)70230-9

Technique

Endoscopic retrograde cholangiopancreatography (ERCP).

Materials

- Duodenoscope: ED450XT8, Fujinon Inc, Saitama.
- Wire: Hydrophilic glidewire, Terumo, Tokyo, Japan; Biliary wire guide, Cook Medical, Winston-Salem, NC, USA.
- Balloon catheter: Cook Medical, Winston-Salem, NC, USA.
- Dilation catheter: 4 mm width, 30 mm length, Cook Medical, Winston-Salem, NC, USA.

Background and Endoscopic Procedure

A 37-year old woman with primary sclerosing cholangitis (PSC) was admitted for ERCP because of pruritus and abdominal pain with moderately elevated biochemical parameters. Before ERCP was performed, an antibiotic prophylaxis was administered. This is an important measure before any endoscopic intervention in PSC, because the majority of patients have colonization with gram-negative bacteria within the biliary tree, and are therefore at high risk of procedure-related sepsis.^{1,2} Aside from this, the overall

complication rate after therapeutic ERCP in patients with PSC is similar to that in patients without PSC.³

At ERCP, the biliary tree is best explored with a balloon catheter to obtain an occlusion cholangiogram of the different biliary segments. This technique allows contrasting the biliary tree with gentle pressure by preventing distal flow of the contrast medium and overcome the resistance of the sclerotized intrahepatic bile ducts. However, it is important to limit contrast injection to an absolute minimum to avoid postprocedural cholangitis. The typical appearance of PSC at ERCP is nearly pathognomonic: multifocal strictures, beading and thinning of the intrahepatic bile ducts, and areas of dilation. There is a strong association between PSC and the development of cholangiocellular carcinoma. These desmoplastic carcinomas are extremely difficult to visualize as they mostly appear just as simple PSC strictures. This is true for all imaging techniques including ERCP, computed tomography scan, and magnetic resonance imaging. Endoscopic brushing and other tissue sampling methods are therefore important, although their sensitivity is suboptimal (50–60%). At ERCP there are two clues that might indicate the onset of malignancy: (1) Identification of a new or modified stricture; and (2) dilation of the bile duct proximal to an extrahepatic stricture. As sclerosing cholangitis is a chronic fibrosing condition, it is difficult for the bile ducts to dilate unless there is a significant pressure gradient generated by obstructions. Therefore, new bile duct dilation is suggestive of the development of malignancy.⁴

Although there is no evidence that endoscopic therapy slows the progression of liver disease, in selected situations it is likely to reduce the risk of cholestasis and infectious complications and offer the patient symptomatic improvement. Which strictures are amenable to endoscopic therapy? At the level of the common bile duct endoscopic dilation allows opening of short- and long-segment stenoses. In contrast, intrahepatic strictures are mostly multifocal and therefore rarely amenable to endoscopic therapy. However, in the hepatic ducts near to

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the bifurcation, short-segment stenoses (< 10 mm) also may be treated endoscopically, whereas long-segment stenoses mostly do not respond to dilation.⁵ This is demonstrated in the present case where a long-segment stenosis in the left hepatic duct is treated. After dilation, there is no sustainable response to treatment. Nevertheless, stent placement should be avoided because of the risk of bacterial overgrowth.

In the present case, after dilation treatment the patient had improvement of clinical symptoms, especially pruritus, but no improvement of biochemical parameters.

Key Learning Points/Tips and Tricks

- Before any endoscopic intervention in PSC, always administer antibiotic prophylaxis against gram-negative bacteria to prevent procedure-related sepsis.
- Endoscopic dilation allows the opening of short- and long-segment stenoses of the common bile duct. Short- but not long-segment stenoses of the hepatic ducts near to the bifurcation also may be treated endoscopically.
- New bile duct dilation may be a clue to the onset of malignancy.
- Stent placement should be avoided because of the risk of bacterial overgrowth subsequent to biliary infection.

Scripted Voiceover

Here we perform ERCP on a 37-year old lady with known PSC. She was admitted because of pruritus, and elevated biochemical parameters. Remember: before we start any endoscopic intervention in PSC we should administer antibiotic prophylaxis to prevent procedure related sepsis. In PSC the biliary tree is best explored with a balloon catheter to obtain a selected occlusion cholangiogram of the different biliary segments. This is because the high resistance of intrahepatic fibrotic stenoses impairs filling with contrast medium.

After half-way inflation of the balloon at the level of the bifurcation we contrast the biliary tree with gentle pressure. Keep in mind to limit contrast injection to an absolute minimum to avoid postprocedural cholangitis. During injection the catheter is slowly pulled back to allow filling of the common bile duct. This contrasted curved duct is the cystic duct, that branches off the common bile duct at an unusually high level. Look: Without inflation of the balloon large

amounts of contrast medium just flow back into the common hepatic duct and eventually fill the gall bladder. This is because of the high resistance of the sclerotic bile ducts in PSC. After inflation of the balloon an occlusion cholangiogram of the right hepatic duct reveals the typical features of PSC that are nearly pathognomonic: we see multifocal strictures, focal dilations and thinning of the intrahepatic bile ducts. Have a close look at the biliary tree: We see multiple strictures but also a paucity of intrahepatic bile ducts with typical thinning in the periphery. During slow withdrawal of the half-way blocked balloon catheter we identify the left hepatic duct with a long-segment stenosis. This dominant stricture has a length of 25 mm and appears somehow distorted and destructed. It is virtually impossible to distinguish between a malignant stenosis and non-neoplastic PSC strictures as they have the same macroscopic appearance. While short intrahepatic stenoses are good indications for dilation strictures with a length of more than 15 mm have a rather poor outcome after endoscopic treatment. However, considering the new onset of clinical symptoms we give it a try. After exchange of the hydrophilic guide wire with a biliary wire guide a dilation balloon with 30 mm length is inserted. The markings on the balloon catheter identify the proximal and distal ends of the balloon. The balloon is positioned right within the stricture and inflated to 4 mm. The balloon is positioned right within the stricture and inflated to 4 mm. The stenosis is indeed very tight during dilation for 60 s. Here we see the contrast medium within the balloon. After dilation repeated contrast injection shows no verifiable response to treatment although the stricture appears more smooth. At this point we decide to finish the procedure today and proceed to dilation up to 6 mm if clinical symptoms do not improve during follow-up.

References

1. Stiehl, A. Primary Sclerosing Cholangitis: The Role of Endoscopic Therapy. *Semin. Liver. Dis.* **2006**, *26*, 62–68.
2. Pohl, J.; Ring, A.; Stremmel, W.; Stiehl, A. The Role of Dominant Stenoses in Bacterial Infections of Bile Ducts in Primary Sclerosing Cholangitis. *Eur. J. Gastroenterol. Hepatol.* **2006**, *18*, 69–74.
3. Etzel, J. P.; Eng, S. C.; Ko, C. W.; *et al.* Complications After ERCP in Patients with Primary Sclerosing Cholangitis. *Gastrointest. Endosc.* **2008**, *67*(4), 643–648.
4. Classen, M.; Tytgat, G. N. J.; Lightdale, C. J. *Cholangiocellular Carcinoma*; Thieme: New York, **2002**; pp 626–627.
5. EASL. Clinical Practice Guidelines of Cholestatic Liver Diseases. *J. Hepatol.* **2009**, *51*, 237–267.